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ABSTRACT

This booklet presents information regarding the history, incidence, and effects of Down Syndrome. The first chapter, presenting an historical perspective of the condition, provides information on counseling parents of Down Syndrome children, and the chromosome structures seen in Down Syndrome patients. The next chapter discusses medical aspects in Down Syndrome, including associated conditions such as visual and hearing disorders, immunological problems, thyroid disorders, congenital heart disease, intestinal disorders, skeletal abnormalities, and use of facial plastic surgery to alter facial features of Down Syndrome patients. A chapter on the treatment of persons with Down Syndrome considers effects of drug treatments. Educational strategies are discussed in the fourth section. The final two sections consider the world of work and living arrangements for individuals with Down Syndrome. (CB)

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AN OVERVIEW OF DOWN SYNDROME

Siegfried M. Pueschel, M.D., Ph.D., M.P.H.

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INTRODUCTION TO THE ASSOCIATION FOR RETARDED CITIZENS OF THE UNITED STATES

Children and adults with mental retardation, their families and friends as well as the professionals that serve them are the Association for Retarded Citizens of the United States. These people, located throughout the United States, function in local and state chapters along with a national office solely devoted to:

- Creating, providing, improving and expanding education, recreation, health, occupational and other programs for persons with mental retardation.
- Supporting research and other programs regarding the prevention and cure of mental retardation.
- Developing and distributing information to improve understanding of mental retardation.
- Advocating for appropriate public policy to insure that persons with mental retardation access their rights as citizens.

Since its inception in 1950, the ARC has operated across the country to improve the welfare of children and adults with mental retardation and their families.

New members are always welcome and needed to expand the strength and influence of the ARC movement. Join your local ARC chapter and become a part of our work. For information about the location of your local chapter contact:

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A Historical Perspective

Although John Langdon Down provided in 1866 an accurate description of the condition that bears his name, historical evidence of Down syndrome goes back much further in time. Early records stem from excavations of a Saxon skull in the 7th century, which is thought to be from a child with Down syndrome. There are also reports of some early paintings from the 16th and 17th centuries depicting children with features as seen in Down syndrome.

The first description of a child presumed to have had Down syndrome was given by Esquirol in 1838. Eight years later, Seguin described a child with characteristics resembling this syndrome, and called it "furfuraceous idiocy." In 1866, Duncan noted a girl "with a small round head, Chinese-looking eyes, projecting a large tongue who only knew a few words." That same year, Down published a paper describing some of the characteristics of this syndrome:

> "The hair is not black as in the real Mongol, but of a brownish colour, straight and scanty. The face is flat and broad, and destitute of prominence. The cheeks are roundish and extended laterally. The eyes are obliquely placed and the internal canthi more than normally distant from one another. The palpebral fissure is very narrow . . . the lips are large and thick and with transverse fissures. The tongue is long, thick, and is much roughened. The nose is small."

Down's great contribution was his recognition of the physical features and his description of the condition as a distinct and separate entity.

Along with many other contemporary scientists of the mid-19th century, Down was influenced by Charles Darwin's book, The Origin of Species. In keeping with Darwin's theory of evolution, Down believed that this condition presented a reversion to a primitive racial type. Recognizing what he believed to be a somewhat Oriental appearance in individuals he observed, Down coined the term "mongolism," and inappropriately called the condition "Mongolian Idiocy."

Today, we know that the racial implications are incorrect. For this reason, but also because of the negative connotation and the derogatory and dehumanizing implications of the terms "mongol," "mongoloid," and "mongolism," such terminology should not be used.

Subsequent to Down's report, many articles have appeared in the literature describing additional abnormal physical findings in Down syndrome. Moreover, speculations relating to the cause of Down syndrome and various treatment approaches have been discussed in the medical literature.

Counseling of Parents

Over the last few decades, the approach to counseling parents has changed dramatically. In the past, physicians, as well as other professionals, often recommended residential placement when a child with Down syndrome was born. Since they felt that the child "will have to leave the family sooner or later," proponents of early institutionalization often argued that attachment of the mother to the child could be prevented by disallowing the maternal instincts to surface. The child with Down syndrome was usually presented to parents as a severely or profoundly retarded infant who "will never do anything," "will be a menace to society," "will become a destructive force within the family,"

or "will have a negative influence on brothers and sisters." Today we know that these assumptions are incorrect. We are aware that institutionalization will have a detrimental effect on the child's development. Unless serious medical conditions exist, children with Down syndrome should be nurtured in the home environment. Most physicians now recognize that in order to maximally develop and mature, every child, including the child with Down syndrome, needs love, affection, and security — and the family is usually the most appropriate source for meeting these needs. In case the biological parents — for whatever reason — cannot keep the child, foster care or adoption are realistic alternatives.

After the birth of a child with Down syndrome, it is of utmost importance that parents be approached by professionals with tact, compassion, and truthfulness. The way parents are counseled during this initial time period will have a vital influence on their subsequent adjustment. If parents are approached in a supportive and positive manner, and if they are told that their infant, although having Down syndrome is first and foremost a human being, then they also will see positive attributes in their infant and their profound distress will be lessened. During this initial period of adjustment, the parents' general human and parental competence should be stressed. The critical role of parenthood needs emphasis, as does the need to provide the infant with a chance to be nurtured and loved.

While professionals should explain to both parents the meaning of Down syndrome, the anticipated development of the child, the chromosomal problem and related issues, other parents who have an older child with Down syndrome can also be helpful. These parents, who have been through similar emotional stress when their child was born, are often more sensitive and can assist the new parents to adjust and to cope more effectively. They are living proof that one can

survive such a crisis, and they can communicate to the new parents that there can be true happiness in the family with a child who has Down syndrome.

A number of Parent-to-Parent Programs have been established throughout the country to help new parents in their distress. For example, a Parent Resource Program has been established in Rhode Island. Newborn nurseries throughout the state maintain a list of names of parents of older children with Down syndrome who are willing to go to the hospital and talk to the new parents when a child with Down syndrome is born. While initially some professionals were skeptical (e.g., anticipating that experienced parents might give the new parents a much "too optimistic outlook"), they soon found out that the resource parents can indeed assist the new parents in their coping and adjustment process in a way the professional is unable to do.

Investigations[2] of parents who had been counseled appropriately after their child with Down syndrome was born revealed that those who were able to cope and adjust to the situation, provided better care and stimulation to the infant, which in turn resulted in accelerated development of the child. Therefore, it should be emphasized that genuine support and positive counseling offered to parents after the birth of their child with Down syndrome is of critical importance for the future quality of life of the child and his family.

Chromosomes in Down Syndrome

In the early 1930s, some physicians suspected a chromosome problem to be present in persons with Down syndrome. At that time, however, the technology of examining chromosomes was not advanced enough to confirm this suspicion.

Only in the mid-1950s did new methods to study chro-

mosomes become available, which in 1959 led to the discovery by Lejeune, who found that the child with Down syndrome has one extra small chromosome. He noted 47 chromosomes in each cell instead of the normal number of 46 chromosomes; instead of the usually observed two #21 chromosomes, he found three #21 chromosomes in each cell which led to the term trisomy 21.

As shown in Figure 1, the chromosomes are arranged in pairs. However, as indicated by the arrow there are three chromosomes in the #21 group in Down syndrome. It is known that approximately 95 percent of children with Down syndrome have this chromosomal condition, trisomy 21.

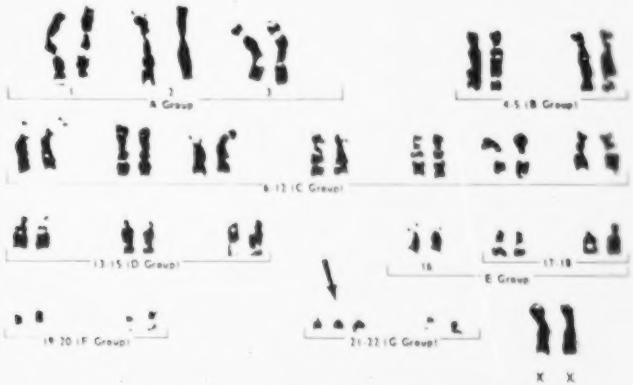

Fig. 1 These are the chromosomes (karyotype) of a girl with Down syndrome. Note the extra # 21 chromosome as shown by the arrow.

It is important to realize that the very first cell resulting from the union of sperm and egg has 47 chromosomes. The extra chromosome can be contributed either by the egg or the sperm, as seen in Figure 2 (See page 6). Every cell derived from this initial cell will likewise have 47 chromosomes and, if not miscarried, a child with Down syndrome will be born.

It has been estimated that approximately 80% of conceptions with trisomy 21 will be miscarried.

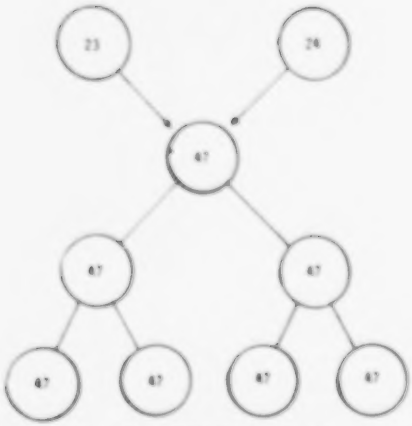

Fig 2. Cell division in Trisomy 21.

There are two other chromosome problems observed in children with Down syndrome. In approximately four to five percent of these children, the extra #21 chromosome is attached to another chromosome, usually to chromosome #14, #21, or #22. This condition is called "translocation." In this instance, the total count of chromosomes will be 46, because two chromosomes are attached to each other, as indicated in Figure 3.

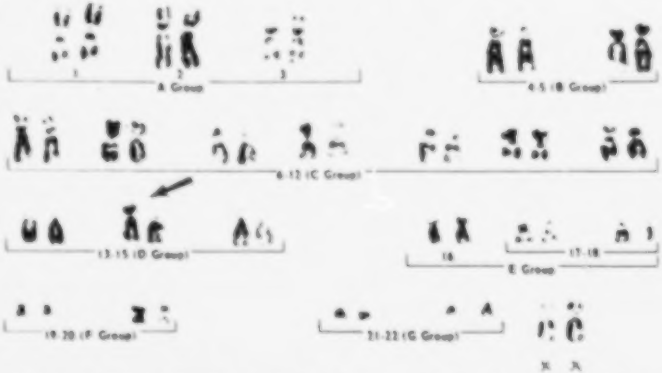

Fig. 3 This is a karotype of a girl with translocation Down syndrome. The arrow indicates the extra #21 chromosome which is "translocated" or attached to a #14 chromosome.

It is important to find out whether or not a child with this type of Down syndrome (translocation) has a parent who is a "carrier." If one of the parents is a carrier, there is an increased risk that this parent will have more children with Down syndrome. Parents are then in need of specific genetic counseling. Furthermore, there is an increased risk that their unaffected children may be carriers of the translocation and have offspring with this syndrome.

The third type of chromosome problem in children with Down syndrome is called "mosaicism." It is least common, occurring in approximately one percent of children with this syndrome. Mosaicism is thought to be due to an error in one of the initial cell divisions shortly after conception, as noted in Figure 4. When the baby is born, one finds some cells with 47 chromosomes and other cells with the normal number of 46 chromosomes which gives a "kind of mosaic picture," therefore the term "mosaicism."

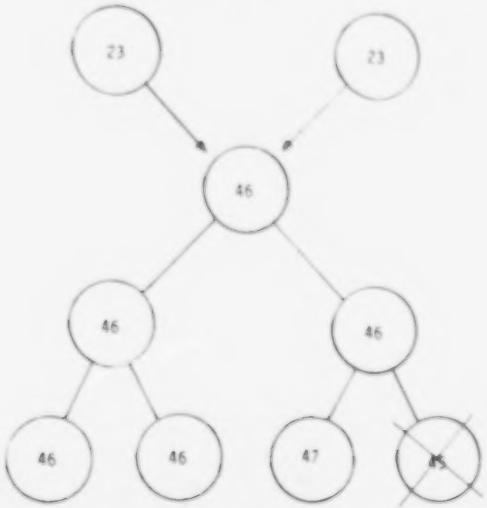

Fig. 4 In mosaicism the "accident of nature" (nondisjunction) is thought to occur during one of the early cell divisions. When this infant is born one will find some cells with 46 chromosomes and others with 47 chromosomes. Cells with 45 or less chromosomes usually do not survive.

Regardless of whether the child has trisomy 21, translocation or mosaicism, the extra chromosome #21 is always associated with specific physical features and mental retardation observed in the majority of children with Down syndrome.

During the past decades, new methods in visualizing details of individual chromosomes allowed the identification of a few children who only had parts of additional #21 chromosome material. Using these new "banding" techniques, it was found that it is not the entire extra chromosome #21, but only a small segment of it that causes the condition. It is not known, however, in what way this extra chromosome material interferes with the development of the unborn child or how the extra genes lead to abnormal brain function.

During the past century many investigators have speculated about the cause of Down syndrome. While the reasons that chromosomes do not divide properly is still not understood (See Figure 5), new theories of causation have been developed in recent years. It has been postulated that radiation and x-ray exposure, viral infections, certain potent drugs, hormonal or immunologic problems may be responsible for the chromosome anomaly in Down syndrome.

Previously, mothers were often blamed for bringing children with Down syndrome into the world, since it is well known that there is a relationship between maternal age and incidence of Down syndrome i.e., the older the mother the greater the risk of giving birth to a child with Down syndrome. Recent reports, however, indicate that there may also be a relationship between the father's age and the incidence of Down syndrome. In addition, studies done during the past decades which examined chromosomes from both parents found that in 20 percent to 30 percent of cases, the extra chromosome #21 comes from the father.

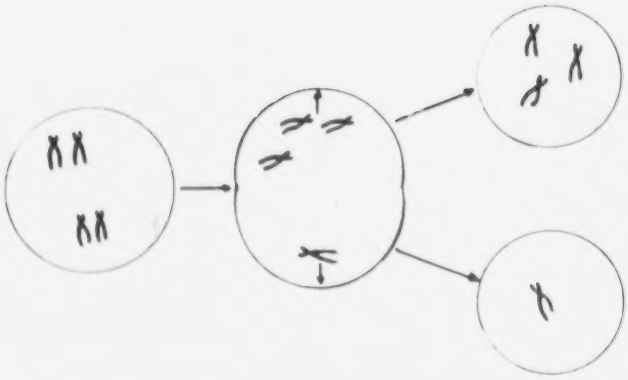

Fig. 5 During the process of cell division, two #21 chromosomes "stick together" (nondisjunction). In the following cell generation one cell will have one chromosome less (this is not a viable cell) and the other cell will have one additional chromosome. For demonstration purposes only, two pairs of chromosomes are shown here.

Medical Aspects in Down Syndrome

There are many organs within the body of a person with Down syndrome that may be adversely affected. In the following discussion only a few selected body organs and systems will be described.

Visual Disorders. There are numerous eye problems associated with Down syndrome. Abnormalities have been uncovered in almost every part of the eye. It has been found that 30 percent of persons with Down syndrome have strabismus (cross-eyedness) which is often due to myopia (nearsightedness). Nystagmus (rapid eye movements) is another frequently observed finding in children with Down syndrome. Approximately 15 percent to 20 percent of these children have nystagmus. Cataracts (cloudiness of the lens) are also noted more often in Down syndrome than in the general population. While congenital cataracts are rare (approximately two percent to three percent), acquired

cataracts have been observed in 30 percent to 60 percent of adults with Down syndrome. Apparently, these cataracts seldom become so dense that they warrant surgical removal. Myopia (nearsightedness) and hyperopia (farsightedness) are also common. It has been estimated that more than 50 percent to 70 percent of children with Down syndrome have to wear glasses because of visual impairments.

Since eye problems are observed frequently, persons with Down syndrome should have periodic eye examinations by a competent ophthalmologist who is able and willing to fully examine and treat (if necessary) persons with handicapping conditions. Many of the eye problems occuring in individuals with Down syndrome can be improved or corrected.

Hearing Impairments. There have been numerous reports in the literature discussing hearing difficulties in persons with Down syndrome. It has been estimated that up to 80 percent of young children with Down syndrome have a mild to moderate hearing loss. Most often the hearing loss is due to conductive problems, and it is less frequently sensorineural in origin. Accumulation of fluid in the middle ear occurs often in children with Down syndrome, which may result in a mild, conductive hearing loss. Hearing deficits may also be caused by an abnormality of the small bones in the middle ear which transmit the sound waves from the eardrum to the inner ear.

If a hearing problem has been identified, appropriate treatment should be initiated. Should the hearing deficit be due to chronic fluid accumulation within the middle ear, tubes can be inserted into the eardrum to drain this fluid. If the hearing deficit is severe, children with Down syndrome may benefit from wearing a hearing aid.

Both optimal visual function and adequate hearing are of utmost importance for any child. However, if the child is mentally retarded, the additional disability of a sensory impairment may further handicap the child, interfering with learning. Therefore, regular hearing assessments and visual examinations should be carried out.

Immunological Problems. The human body has specific defense mechanisms which render it resistant to certain diseases. There are special organs in the body (e.g., thymus gland and bone marrow) producing specific cells that provide protection against invading germs. Some of these special cells are discharged into the blood stream and, at strategic parts of the body, they literally trap and digest bacteria and other microorganisms. Other special blood cells (lymphocytes and plasma cells) produce certain substances which are important in the body's immune system. In addition, our skin provides chemical and cellular defenses which ordinarily do not allow germs to invade our body. The intact immune system is able to recognize, destroy and eliminate foreign substances.

Frequently the question has been raised, "does the child with Down syndrome have appropriate protection and resistance against bacterial and other foreign invaders?" Unfortunately, there is no clear-cut answer to this question. A study of the pertinent medical literature on this issue reveals that some investigators have found minor defects in one or the other system of immunity, while other researchers have not uncovered such deficiencies. A number of subtle changes in the defense mechanism of the body in persons with Down syndrome have been reported, however, most children with Down syndrome do not have serious problems with their immune system.

While the thymus gland (which is located beneath the

upper part of the breastbone) is of normal size, structural abnormalities of the thymus gland have been noted in some children with Down syndrome. There is also a lack of certain cells (T-lymphocytes) in the thymus which are of great importance in the overall defense strategies of the body. In addition, there are fewer of these cells in the circulating blood of children with Down syndrome. Yet, the slightly reduced number of cells does not cause severe ill effects in the child. However, the fact that these cells function abnormally may be in part responsible for the increased occurrence of infections reported by some investigators. Other studies[2] did not observe significantly more infectious problems in young children with Down syndrome than in normal children of similar age.

Researchers have also reported reduced numbers of other blood cells (B-lymphocytes). These cells produce special proteins (gamma globulins) which are an important component of the general immune system. Some observers have reported a decrease of these proteins in the blood of children with Down syndrome, while other researchers have reported an increase. Most of the so-called immune globulins, however, are within the normal range.

Another important immunologic function relates to specific antibody production in response to such immunizations as tetanus, diphtheria, measles, polio, mumps and others. Most children with Down syndrome respond in the same way to these vaccines as do other children without chromosomal disorders. However, antibody production to other antigens has been reported to be impaired in older persons with Down syndrome.

Because of the increase of respiratory infections observed by some investigators, the function of certain white blood cells (leukocytes) has been studied in children with Down syndrome. Some studies found that these cells

respond to infections in a normal fashion, while others reported a limited ability of these cells to ingest or kill specific germs. It should be noted that a number of individuals with Down syndrome frequently have skin infections which may result in recurrent formation of absesses, primarily in the area of the buttocks and thighs.

The increased prevalence of leukemia in children with Down syndrome which is 7 to 20 times higher than in the normal population, also has been linked to a possible defect in the immune mechanisms. Vitamin A, Vitamin E, zinc and a variety of other substances have been found to have some influence on the immune system in persons with Down syndrome. More research is needed in this area, since studies to date have often yielded conflicting results.

Thyroid Disorders. Many reports in the literature discuss thyroid dysfunction in Down syndrome. Although most children with this chromosome disorder have normal thyroid function, there is an increased incidence of hypothyroidism (low levels of thyroid hormone) in persons with Down syndrome. Depending on the age group studied, the incidence of thyroid problems varies from 3 percent to 27 percent. Hypothyroidism is more often found in the adult with Down syndrome. Thyroid autoantibodies (antibodies against their own thyroid gland) have also been frequently uncovered in individuals with Down syndrome.

The thyroid gland has important functions within the human body. If an inadequate amount of thyroid hormone is available, the young child's intellectual function will be adversely affected. Therefore, it is important to examine regularly children's thyroid function in order to prevent further brain damage. Since children with inadequate thyroid hormone production will have stunted growth, regular

growth measurements should be made. If significant retardation in growth is observed, thyroid function studies should be carried out. If a child with Down syndrome is found to have a thryoid disorder, appropriate treatment should be forthcoming, in order to optimize his or her growth and development, learning ability, and vocational achievement.

Congenital Heart Disease. Congenital heart disease is observed in 30 percent to 40 percent of children with Down syndrome. At times, the heart defect may be a minor problem if there is only a small hole in the wall between the heart chambers. In other situations the heart defect might be so severe that it may be life threatening. In particular, in some children with complex heart problems, such as complete atrio-ventricular canal or endocardial cushion defect, and tetraloy of Fallot, the valves are defective and holes are found between the different chambers. Prior to recent medical advances, many children with these severe congenital heart defects died.

Years ago, cardiologists and cardiac surgeons often were relunctant to recommend or to perform heart surgery on children with Down syndrome. These physicians often felt that children with Down syndrome were so severely intellectually handicapped that surgical intervention would not benefit the child in the long run. Other arguments against cardiac surgery often stressed reduced life expectancy, unemployability, and financial considerations.

During recent years, however, technologic advances in cardiovascular surgery, as well as society's altered viewpoint concerning handicapped persons, have brought about changes in the management of congenital heart disease in individuals with Down syndrome. Cardiologists and cardiac surgeons recognize that many individuals with Down syndrome can lead meaningful lives and that they should be treated as other children. Appropriate medical

management, including the administration of digitalis and other drugs (diuretics) if indicated, and prompt surgical repair at the optimal time will improve the quality of life of the individual with Down syndrome.

Intestinal disorders. In general, significant intestinal problems are not common in Down syndrome. There is, however, a higher incidence of duodenal obstruction (blockage of the segment of intestine which is adjacent to the stomach). This blockage prevents food from passing through the intestinal tract causing death from starvation if prompt surgical repair is not done. In previous years, children with Down syndrome frequently were not operated when they had such a blockage, since some professionals felt that it was "undesirable" to keep them alive. Fortunately, such an inhumane and unethical attitude does generally not prevail at this day and age. Of course, surgery should be performed when an intestinal obstruction is diagnosed in the newborn child and no medical treatment of any kind should be withheld from a child with Down syndrome that would be given unhesitatingly to a child without this chromosomal disorder.

Since neurologic problems in the child with Down syndrome, including poor muscle tone (muscle weakness) and poor sucking, may cause feeding difficulties, special attention should be given to the young child's nutrition. Some of these children also tend to spit up more than normal infants do, but with thickened feedings and maintaining the infant in a half-upright position after meals, this tendency can be overcome.

Constipation is more often observed in small children with Down syndrome than in infants who do not have this chromosome disorder. This problem can usually be overcome by adding fruit and juices to the child's diet. Laxatives are rarely needed.

Obesity is often observed in many older persons with Down syndrome. It is the result of both excessive caloric intake and decreased exercise. Therefore, it is important to establish appropriate dietary habits from early life onward. Starchy and fatty foods, as well as sweets, should be avoided, and a regular exercise program should be an integral part of the individual's life.

Skeletal Abnormalities. Skeletal problems in Down syndrome are common, and may be found in many parts of the body. The skull is usually small, and some of the facial bones are hypoplastic (reduced in size). Rib abnormalities and a structurally altered pelvis are frequently noted. Occasionally, hip or kneecap dislocations are observed. Many children show hyperflexibility at the joints (double jointedness), but this does not usually interfere with normal movement.

One skeletal condition called "atlanto-axial instability" is observed in 15 to 20 percent of persons with Down syndrome. It occurs because of the laxity of the ligaments which hold the joints of the first two vertebrae of the neck together. If the nerves of the spinal column are damaged because of abnormal movements of these neck bones, the child may have problems with walking, may complain of discomfort in the neck area, and have other neurologic symptoms. The latter condition, also called symptomatic atlanto-axial subluxation occurs in about one to two percent of individuals with Down syndrome and these patients will have to undergo surgery to correct this neck problem. Persons with Down syndrome who want to participate in athletic activities (such as in Special Olympics) should first be examined by a physician and have X-rays of their neck to determine if this condition is or is not present. If individuals with Down syndrome have atlanto-axial instability they will not be able to participate in those sports activities that potentially could injure their neck.

Facial Plastic Surgery. Recently, surgery has been advocated to alter some of the physical features commonly observed in persons with Down syndrome. Some plastic surgeons feel that if the appearance of these children can be improved by removing skin folds between the eyes and the nose (epicanthal folds), raising the bridge of the nose, correcting the receding chin and reducing the size of the tongue, they may become more "socially acceptable." Such surgical intervention is controversial, because many people feel it is unnecessary.

For further information on other medical aspects in persons with Down syndrome, the reader is refered to more comprehensive texts.[3-4]

Treatment of Persons with Down Syndrome

During the past decades a multitude of treatments for individuals with Down syndrome have been reported in the literature. A variety of medications, including hormones, vitamins, sicca cells, dimethylsulfoxide, and various combinations thereof have been employed in an attempt to improve physical features and mental functioning in persons with Down syndrome.

Probably the first reported drug treatment was attempted at the end of the last century when thyroid hormone was given to children with Down syndrome. Since then, many physicians have utilized thyroid hormone therapy in persons with Down syndrome, and many professionals still use such treatment today. A controlled study in which a group of children receiving thyroid hormone was compared with a group receiving a placebo reported no significant difference in the overall function of the two groups.[5]

Pituitary extract also has been employed in the treat-

ment of children with Down syndrome. While claims have been made of marked improvement, the current consensus is that pituitary extract does not benefit these children's intellectual and social developments.

Glutamic acid and its derivatives have been used for children with Down syndrome since the 1940s. Many workers initially reported positive results, but more recent studies have not supported those early conclusions.

Dimethylsulfoxide also has been given to children with Down syndrome. In particular, Chilean investigators have claimed a marked improvement of intellectual functioning in children who had received this chemical. A short-term study done in Oregon, however, did not reveal any significant positive changes in chidren with Down syndrome who were given dimethylsulfoxide.

Sicca cell or dry cell treatment has been advocated in Europe for children with Down syndrome during the past few decades. Sicca cell therapy consists of injections of cells prepared from fetal animal organs. Such treatment allegedly stimulates the growth and function of the corresponding tissues in the human body. A recent West Coast publication has brought this form of treatment into the limelight on this continent. Reports from the European scientific literature, as well as results from a study done in Canada, however, do not support the claims made by proponents of this therapy. Recent scientific publications suggest that animal tissue injected into human beings may produce a "slow virus disease" many years later.

A variety of medications, including minerals, vitamins, enzymes, hormones, and other substances, referred to as the U-series, have been used by Turkel[6] for treatment of children with Down syndrome. Although improvement has been reported by Turkel and some Japanese physicians who used the U-series, a double blind study carried out in

1964 did not uncover any significant difference in intellectual function between children given the U-series and others who had been administered a placebo.

A 1981 article claimed that three of four children with Down syndrome had gains of 10 to 25 IQ points when given large amounts of 11 vitamins and 8 minerals. Reduction of the physical characteristics of Down syndrome were also reported. Unfortunately, the study design was rather poor. There was no random assignment of children to study and control groups, unequal distribution between study and control groups, failure to control for seizures, unusual behaviors or sensory impairments, use of three different psychological tests, and use of inappropriate test procedures to study thyroid function in these children. Subsequent studies that attempted to replicate this study using the same "treatment" approach, did not find any beneficial effects.

Since it was reported in the mid-1960s that children with Down syndrome have low blood levels of serotonin, which is an important neurotransmitter, several investigators have administered the precursor of serotonin, 5-hydroxytryptophan (a building stone of protein) in the treatment of this syndrome. Pueschel[2] reported a longitudinal double blind study done at Boston Children's Hospital Medical Center using both 5-hydroxytryptophan and pyridoxine. It was found that these compounds did not have any significant positive effect on motor, social, language and intellectual development of young children with Down syndrome. In spite of the negative results reported by several other investigators using these "medications," some physicians in this country and abroad are still treating children with Down syndrome with 5-hydroxytryptophan and pyridoxine.

In summary, one must say that no drug treatment has been found to be effective to date for children with Down

syndrome. It should be mentioned, however, that recent advances in molecular biology make it feasible now to directly examine the genetic basis of Down syndrome. The challenge for the future is to further isolate, map and characterize gene sequences on chromosome #21 which are responsible for the physical characteristics and the intellectual limitations observed in children with Down syndrome. Identifying the genes and their interference with normal development and counteracting the specific actions of those genes could provide a rational approach to medical therapy in the future.

Educational Strategies

Although no effective medical therapy is currently available for children with Down syndrome, early intervention programs, preschool nurseries and special education programs can substantially foster their development. Considerable evidence indicates that a child's development can be positively influenced by appropriate environmental enrichment. Specifically, early intervention focuses on improving the infant's sensory, motor, and social developments. In recent years, psychologists and educators have generally agreed that it is the quality rather than the sum of total stimulation that shapes the physical and mental developments of the young child. It is, therefore, the structure and the content of an early stimulation program that should be emphasized rather than the indiscriminate use of nonspecific stimuli.

Hayden, a pioneer in developing effective early intervention programs, observed that "Infants with Down syndrome can benefit tremendously from infant learning programs which meet certain basic criteria that are essential for quality program development." The primary goal of such programs is to bring each child as close to developmental norms as possible. To be effective, it is necessary to under-

stand the special needs of the individual child and to have the interest and support of the parents or the caregiver who can maximize the benefits of the child's learning through carryover into the home setting.

At the University of Washington Model Preschool Infant Learning Program,[6] all family members are invited to participate since it is important for all of them to understand the infant with Down syndrome and to respond positively to his or her needs. As with other infants, those with Down syndrome need opportunities to explore their environment. They should be introduced to different objects, to different people, and to different activities that are part of everyday living. Gains made by a child with Down syndrome in such an early intervention program will build the foundation for later learning.

Preschool also plays a very important role in the life of the child by providing social interaction and development of basic self-help skills. Nursery school enables the child to participate in a broader world.

Upon entering school, the child with Down syndrome, like all children, is still growing and maturing. School should provide a stimulating and rich experience, and it should foster feelings of personal identity, self-respect, and enjoyment. The child should be given an opportunity to engage in sharing relationships with others.

Adolescence is frequently a challenging time in the life for both the young person and his family. While this might be a very troublesome time period for those of normal intellectual abilities, the problems of an adolescent with mental retardation are frequently intensified. Many adolescents with Down syndrome may have the physical attributes of normal youngsters, yet they often do not possess the intellectual or behavioral capabilities to cope with either the demands of

their environment or their own desire for independence. They are faced with preparing for vocational competence as well as developing an array of social skills needed to function in society.

The World of Work

A vocational program for young persons with Down syndrome should be designed to provide them with a structured and realistic vocational experience that will enable the student to achieve maximum success in either community employment or sheltered work. Such a program must be based on the vocational needs of the students, and should emphasize the development of interpersonal skills required in most jobs.

Success of the individual with Down syndrome in the job market is, in part, based on his or her efficiency in doing a job, and coupled with such traits as reliability, diligence, responsibility, and trustworthiness on the job. Once on the job, persons with Down syndrome will need to establish a meaningful relationship with their employer and co-workers. They often impress their employers with skills and attitudes that they are not able to verbalize when initially seeking employment.

Undoubtedly, the first few days and weeks of adjustment on the job are critical. In some instances, frustrations faced in the new environment could result in the loss of the job. Therefore, during this initial adjustment, assistance is needed to guide the young person with Down syndrome in order to avoid failure and prevent a premature withdrawal from a job situation.

Too frequently, handicapped individuals, including persons with Down syndrome, are still vocationally stereotyped. They are often ushered into positions such as

dishwashers and janitors. While these occupations might be complex and might be personally satisfying and rewarding to many individuals, they often present a lack of consideration for the individual worker's interest and aptitude. Yet, creativity and imagination can open up a wider variety of occupations for the handicapped person.

Whether the employment is in private industry or in a sheltered workshop, if appropriately placed, a person with Down syndrome will gain a feeling of self-worth and of making a contribution to society. This experience is important for every person, but in particular for the handicapped individual. The person with Down syndrome should be given the opportunity to prove to himself and to others that he is capable of accomplishing vocational tasks since such experiences will greatly increase his self-esteem.

Living Arrangements

The individual with Down syndrome who has finished school and job training may leave home just like other individuals to form new living arrangements. In planning community living for the adult with Down syndrome, one has to determine the individual needs of the person and then see whether the community is able to meet these needs.

Failure to satisfy both physiological and psychological needs may lead to unsuccessful community living. It is important that one considers these needs, since adults with Down syndrome have often been treated as though their only problem was mental retardation. Adults with Down syndrome living in the community should be provided with services that are broad and comprehensive. One should take into consideration such factors as age, sex, degree of mental handicap, medical and emotional problems, job arrangements, recreational opportunities, and the availability of transportation It is important that living quarters not only be

designed in terms of bed space, but also take into account the total needs of the handicapped person. There may be homes where adults with Down syndrome live in near total independence, there may be others where parents or responsible relatives are in charge, or halfway houses with considerable ongoing supervision and available supportive services. Others may choose to live in apartments where varying degrees of supervision and assistance are available from staff.

Community programs should enable adults with Down syndrome to perform adequately the activities of daily living, to live harmoniously in the community, to achieve the maximum level of productivity, and to fulfill normal civic responsibilities according to their abilities. In addition to vocational experiences, there should be time for social interaction and recreational activities.

While the planning for adults with Down syndrome in the community might rest with a particular agency, it is important to involve in such planning those who receive services. As adults with Down syndrome develop maturity and social competence, they will demonstrate capabilities for participating in self-direction and decision-making that could contribute to their own independent living in the community, which should make their life more meaningful.

In order to accomplish these goals, attitudes will have to be forged in society so that young men and women with Down syndrome will be accepted for what they are so that they may, to the best of their abilities, realistically participate in community life.

Conclusions

While there have been many significant advances during the past decades concerning Down syndrome, there is room for improvement and progress, and undoubtedly major challenges are ahead of us.

It is hoped that in the future, persons wth Down syndrome will be offered a status that observes their rights and privileges as citizens in a democratic society and, in a real sense, preserves their human dignity. Society has to realize that persons with mental retardation, including individuals with Down syndrome, are people in their own right, in spite of their limited capacity for academic achievement.

Instead of custodial supervision, community based programs should provide guidance and assistance to help persons with Down syndrome develop their own identities and lead their own lives as much as possible. Such optimism regarding the future will only come true if we retain a solid grip on the reality of the present and pass on the wisdom derived from the past.

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